JACC Vol. 5, No. 6
June 1985:118B–121B

Sudden Unexpected Death in Children and Adolescents

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To determine the incidence and clinicopathologic spectrum of sudden unexpected death, we reviewed the death certificate of all residents of Olmsted County, Minnesota who were between 1 and 22 years of age when they died during the period from January 1950 to October 1982. Of 515 death certificates reviewed, 12 (2.3%) recorded sudden unexpected death, resulting in an incidence of 1.3 per 100,000 patient-years. The subjects ranged in age from 3 to 20 years (median 13); 8 of the 12 were male. Of the 12 deaths, 4 were definitely cardiac-related and 3 were probably cardiac-related. In the five other cases, the cause of death was unknown. Three of the 12 subjects had a history of syncope; 2 of the 3 had syncope associated with exercise, and both died while exercising.

The relative rarity of sudden unexpected death in children and adolescents probably precludes population screening techniques to identify subjects at risk. However, a subset of subjects with 1) exercise-associated syncope, 2) nonvasodepressor syncope, 3) a family history of sudden unexpected death, or 4) a family history of hypertrophic cardiomyopathy deserves extensive and thorough evaluation.

(J Am Coll Cardiol 1985;5:118B–121B)

Ten to 32% of adults die suddenly and unexpectedly (1). This important mode of death has been associated with smoking, coronary artery disease, hypertrophic cardiomyopathy, coronary artery anomalies and arrhythmia. Although sudden unexpected death occurs in children and adolescents, the incidence, to our knowledge, is poorly defined. The purposes of this study were 1) to determine the incidence of sudden unexpected death in children and adolescents, and 2) to develop an approach to identifying the child or adolescent who is at risk for sudden and unexpected death.

Methods

We reviewed the death certificates of all Olmsted County, Minnesota residents who were between 1 and 22 years of age when they died during the period from January 1950 to October 1982. Selecting cases by the cause of death listed on the death certificate, we reviewed the clinic records, hospital records, coroner's reports, autopsy reports and autopsy material (gross heart and cardiac and pulmonary histology) of subjects with obvious or suspected sudden unexpected death or subjects in whom the cause of death was unknown. We defined sudden unexpected death as nontraumatic death occurring instantaneously or within an estimated 24 hours of the onset of acute symptoms or signs (1).

Olmsted County is located in southeastern Minnesota. Except for the city of Rochester, it is a rural community. The major industries are farming, medicine (the Mayo Clinic) and international business machines. The population of Olmsted County, Minnesota is largely middle to upper-middle class. The medical records of all persons living in Olmsted County are maintained at the Mayo Clinic, and all autopsies are performed at the Mayo Clinic.

Results

Incidence. During the 32 year period from 1950 to 1982, 515 Olmsted County residents between 1 and 22 years of age died. Of these 515, 12 (2.3%) died suddenly and unexpectedly. During the same period, there were 930,678 patient-years represented for this age range, resulting in an incidence of 1.3 sudden unexpected deaths per 100,000 patient-years. There was no apparent change in the incidence of sudden unexpected death during the study (Table 1).

Cardiac-related deaths. The 12 subjects (8 male and 4 female) who died suddenly and unexpectedly ranged in age from 3 to 20 years (median 13). The incidence of sudden unexpected death was higher for male subjects (1.7 per 100,000 patient-years) than for female subjects (0.64 per 100,000 patient-years).

Four (33%) of the 12 deaths were definitely cardiac-related and the causes included myocarditis, congenital aortic valve stenosis, hypertrophic cardiomyopathy and primary

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0735-1097/85/$3.30

Table 1. Incidence of Sudden Unexpected Death in Olmsted County, Minnesota, 1950 to 1982*

Year	Patient-Years	Sudden Unexpected Deaths	Incidence (per 100,000)
1950 to 1954	82,859	2	2.4
1955 to 1959	115,663	0	0.0
1960 to 1964	143,253	2	1.4
1965 to 1969	164,315	1	0.6
1970 to 1974	171,941	4	2.3
1975 to 1979	163,309	2	1.2
1980 to 1982	82,486	1	1.2

*All subjects were between 1 and 22 years old at the time of death.

(plexogenic) pulmonary hypertension (Table 2). A 6 year old child with myocarditis and a 3 year old child with hypertrophic cardiomyopathy were siblings and died 10 years apart. A 13 year old boy with congenital aortic valve stenosis had had an aortic valvotomy at 5 years of age and was awaiting an operation because of progressive aortic stenosis with a left ventricular aortic pressure gradient of 88 mm Hg.

Three additional deaths (25%) were probably cardiac-related. A 20 year old woman had subtotal obstruction of the atrioventricular nodal artery, and an 11 year old boy with a history of prior syncopal episodes had histologic evidence of myocardial ischemia predating death by approximately 6 hours. The third patient, a 7 year old boy, had confluent bronchopneumonia at the time of death. However, he was clinically well and died suddenly while riding his bicycle. He had had two documented prior syncopal episodes, and the relation between the bronchopneumonia and his death is unclear.

Cause of death. In 5 (42%) of the 12 deaths, the cause was unknown. One of these patients did not have an autopsy, but review of the autopsy material (heart and lungs) of the other four patients did not reveal an apparent cause of death. Microscopic examination of the cardiac conduction system revealed no abnormalities. The deaths could have been related to arrhythmias.

Three (25%) of the 12 subjects had had prior presyncopal or syncopal episodes, and in 2 the episodes were associated with exercise. An 11 year old boy had been under water for approximately 5 seconds while swimming, after which he surfaced, came to the side of the pool, had a seizure and died. About 1 year before, he had had a similar episode associated with a "dreamlike" state, but this was unassociated with a seizure. A 7 year old boy had a cardiac arrest and died while riding his bicycle. One year previously, as well as 1 week before his death, he had experienced documented exercise-associated syncopal episodes, but the results of a physical examination were normal. A 6 year old girl whose brother had died 10 years before with hypertrophic cardiomyopathy had a syncopal episode 3 weeks before her death; autopsy revealed myocarditis.

Table 2. Summary of 12 Sudden Unexpected Deaths in Olmsted County, Minnesota: Children and Adolescents

Case	Sex	Age at Death (yr)	Year of Death	Cause	Prior Syncope
1	M	16	1950	Unknown	No
2	M	15	1951	Unknown	No
3	M	7	1962	Bronchopneumonia	Yes
4	F	3	1963	PPVOD	No
5	M	20	1968	Unknown	No
6	M	11	1971	Unknown	Yes
7*	M	3	1971	Hypertrophic cardiomyopathy	No
8	F	20	1972	Unknown	No
9	M	13	1974	Aortic stenosis	No
10	F	20	1976	Fibrosis of AV nodal artery	No
11	M	15	1977	Unknown	No
12*	F	6	1981	Myocarditis	Yes

*Siblings. AV = atrioventricular; F = female; M = male; PPVOD = plexogenic pulmonary vascular obstructive disease.

Discussion

Incidence. The incidence and epidemiology of sudden unexpected death in adults have been studied extensively. However, there has been little investigation of this problem in children and adolescents. No doubt this is due to the relative rarity of sudden unexpected death in a young and presumably healthy population. Yet it is a particularly tragic event for a young and vigorous child or adolescent to die suddenly and unexpectedly. In a study of sudden unexpected death in St. Louis County, Missouri, Kennedy and Whitlock (2) found that in 1981 the incidence for children 1 to 9 years old was 2.5 per 100,000 patient-years, whereas in 1982 it was 8.5. The study also showed that in 1981 the incidence for the group 10 to 19 years old was 2.4 per 100,000 patient-years, whereas in 1982 it was 5.3. These values are greater than our incidence of 1.3 per 100,000 patient-years. This difference may be due to a different sampling period (1981 and 1982 in the study by Kennedy and Whitlock compared with 1950 to 1982 in our study) and a markedly different population base. The sampling period is important because the incidence of sudden unexpected death in children and adolescents is variable from year to year. In the study by Kennedy and Whitlock (2), there was a twofold to threefold difference between 1981 and 1982. In our study, the incidence of sudden unexpected death among 5 year periods varied from 0 to 2.4 deaths per 100,000 patient-years. In the series of Kennedy et al. (3), 50% of the patients between 1 and 19 years of age who died suddenly had documented cardiovascular disease, including hypertrophic cardiomyopathy (two patients), congenital heart disease (three patients) and unspecified cardiac disease (three patients).

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Neuspiel and Kuller (4) identified 196 cases of sudden unexpected death in children and adolescents (1 to 21 years of age) in Allegheny County, Pennsylvania that occurred between 1972 and 1983. However, these investigators did not report the incidence of sudden unexpected death. Forty-seven (24%) of the 196 deaths were related to cardiac disease, including myocarditis (14 cases), cardiomyopathy (8 cases), myocardial fibrosis, coronary artery abnormalities, aortic aneurysm, Ebstein's anomaly, prolongation of the QT interval and myocardial infarction. Excluding the patient with myocarditis, 93% of those with cardiac disease were known to have cardiac disease before death.

Possible predictors of sudden death. Although epidemiologic studies of sudden unexpected death in children and adolescents have been limited, there have been numerous isolated reports of patients who have died suddenly and unexpectedly. These reports are biased toward sudden deaths with identifiable causes and deaths from specific entities, such as hypertrophic cardiomyopathy and congenital coronary artery malformations. However, previously published studies allow insight into possible predictors of sudden unexpected death and circumstances surrounding the terminal event. We reviewed 13 studies involving 61 children and adolescents who died suddenly and unexpectedly (5–17). Forty-six of the patients (75%) were male and 9 (15%) were female (in 10%, the gender was not specified). The diagnoses were: hypertrophic cardiomyopathy (57%), anomalous origin of the left coronary artery from the right sinus of Valsalva (24%), aortic valve stenosis (10%), cystic medial necrosis of the aorta (3%) and medial hypertrophy of the sinus node artery (2%); in 3% the cause was unknown. A history of at least one prior syncopal episode was relatively common (29%) for patients with hypertrophic cardiomyopathy, but was found in only 1 (7%) of 15 patients with a coronary artery abnormality, 1 (17%) of 6 patients with aortic valve stenosis and 1 patient with medial hypertrophy of the sinus node artery. Death was associated with exercise or occurred shortly after exercise in 21 (60%) of 35 subjects with hypertrophic cardiomyopathy, in 10 (66%) of 15 subjects with origin of the left coronary artery from the right sinus of Valsalva and in 5 (83%) of 6 subjects with aortic valve stenosis.

Exercise-associated syncope. Three of the 12 subjects in our series had had syncope some time before death, and 2 had had syncope associated with exercise. Because between 12 and 15% of normal adolescents experience vasodepressor syncope (18), syncope itself may not be helpful in identifying the adolescent who is at increased risk for sudden death. However, benign vasodepressor syncope usually occurs in response to emotional stress or pain, in certain environments such as a hot crowded room and while the subject is in an upright or sitting position. It is unusual for vasodepressor syncope to be associated with exercise. Although syncope itself may not be predictive of sudden death, exercise-associated syncope may be, and children and adolescents with exercise-associated syncope should be evaluated thoroughly.

Prevention. Prevention of sudden unexpected death in children and adolescents is dependent on identification of those who are at risk. The relatively low incidence (1.3 to 8.5 per 100,000) of sudden unexpected death in children and adolescents probably precludes mass screening programs utilizing echocardiography or exercise testing to identify those with hypertrophic cardiomyopathy, aortic valve stenosis or coronary artery abnormalities. However, diagnostic screening studies may be appropriate for subsets of children and adolescents who are identified as possibly being at increased risk for sudden unexpected death. We propose that all children and adolescents with 1) exercise-associated syncope, 2) nonvasodepressor syncope, 3) a family history of sudden unexpected death, or 4) a family history of hypertrophic cardiomyopathy have a thorough history and physical examination, a two-dimensional echocardiogram, an exercise electrocardiogram and 24 hour ambulatory electrocardiographic monitoring. Detailed electrophysiologic testing may be indicated in selected instances. Unfortunately, a substantial portion of children and adolescents with sudden unexpected death will not have a prior syncopal episode or family history of sudden unexpected death or hypertrophic cardiomyopathy. Identification of this latter subset before death remains an unsolved problem.

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